Pulmonary Atresia With Ventricular Septal Defect in Adults

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Background Multistage surgery culminating in completed hemodynamic repair is now performed for pulmonary atresia with ventricular septal defect (PA-VSD). Justification for operation in patients with an adequate collateral pulmonary circulation is controversial. Data on natural adult survival are scant but are necessary to provide the rationale for multistage reconstructive procedures.

Methods and Results All cyanotic adults with PA-VSD in the UCLA Adult Congenital Heart Disease Center Registry from 1978 through 1992 formed the basis for this study. Registry data and echocardiographic, hemodynamic, and angiographic information were used to determine longevity, clinical course, and operative feasibility. Of 26 patients, 16 were unoperated when referred (group A), and 10 had been palliated before age 18 years (group B). Two thirds were 18 to 29 years old. Only 2 patients survived beyond age 40 years. Six died during follow-up at a mean age of 31 years (±12.1 SD). Eight group A patients were in New York Heart Association class II, and 8 were in class III. Of the 26 group A and B patients, 20 had aortic regurgitation, which was moderate or severe in 10. Eight had cardiac failure. Of 11 group A patients who remained unoperated, 5 died. Twelve patients were considered eligible for surgery at ≥18 years of age. Ten underwent completed hemodynamic repair with a mean postoperative right ventricular–to–left ventricular systolic pressure ratio of 0.45 (±0.16 SD), and there were no early or late deaths.

Conclusions Even when collateral blood flow permits adult survival, all such patients are symptomatic. Mean life expectancy without operation did not exceed three decades. Aortic regurgitation and cardiac failure are significant negative variables. Nearly half of unoperated adults died during follow-up. Staged hemodynamic repair can be performed with a low surgical risk in properly selected adults with PA/VSD. (Circulation. 1994;89:243-251.)

Key Words • surgery • defects

Staged reconstructive operations are now performed on a broad range of patients with pulmonary atresia and ventricular septal defect (PA-VSD), but the long-term results of complex surgical management remain uncertain. The anatomic variability of the native pulmonary vascular bed and its collateral blood flow in large measure accounts for differences in clinical manifestations, in unoperated survival, and in the complexity of unifocalization operations. The rationale for surgical intervention must be weighed against the mortality and morbidity inherent in the unoperated malformation. The importance of natural survival patterns is twofold: to provide the backdrop against which long-term surgical results can be assessed and to aid in determining the rationale for surgical reconstruction in patients who select themselves out as long-term survivors.

When all patients are considered collectively, life expectancy without surgery can be as low as 50% at 1 year of age and 8% at age 10 years. One end of the spectrum is represented by neonates, infants, and children who experience either severe cyanosis due to inadequate pulmonary blood flow or congestive heart failure due to excessive pulmonary flow. The other end of the spectrum is represented by patients with adequate but not excessive pulmonary blood flow that permits survival into adolescence and adulthood. Previously, little clinical information has been available on the latter group. Between these two extremes are patients who remain clinically stable after palliative procedures but are anatomically suitable for unifocalization repair. For patients who are palliated and stable, the risks and long-term morbidity of multiple, complex surgical procedures must be weighed against the risks inherent in the palliated malformation. The justification for and outcome of surgical management in stable unoperated or stable palliated adolescent and adult survivors remains to be established.

In light of these issues, we sought to determine the prevalence, morbidity, mortality, and surgical status of patients with PA-VSD who reached adulthood. We examined the outcome of unoperated and palliated adult survivors, focusing chiefly on unoperated adults to establish longevity patterns, clinical course, and indications for and feasibility of surgical intervention.

Methods

Between 1978 and 1992, 26 cyanotic patients 18 years of age or older with PA-VSD were referred to the UCLA Adult Congenital Heart Disease Center. Diagnoses were confirmed by cardiac catheterization, echocardiography, or both. Excluded from study were patients with Fallot's tetralogy and acquired PA or with PA occurring in the context of other complex congenital malformations. "PA" as used for this study is defined as the most severe expression of Fallot's tetralogy. A rudimentary pulmonary valve is present but imperforate. The
infundibulum terminates blindly against the atretic valve or against muscle.

Patients were divided into two groups. Group A comprised unoperated adult survivors who reached age 18 years without surgical intervention. Group B comprised adult survivors who were palliated before age 18 years but had not undergone completed hemodynamic repair. "Palliation" refers to any surgical intervention designed to increase pulmonary blood flow without closure of the VSD. Completed hemodynamic repair, achieved through one or more interventions, refers to unifocalizing extraneous aortic-to-pulmonary collaterals, connecting the right ventricle to a native or reconstructed pulmonary artery confluence, and closing the VSD.

The following data were assembled for all patients: incidence within the UCLA Adult Congenital Heart Disease Registry; age distribution; mortality; systemic arterial oxygen saturation; hematocrit, hemoglobin, and red blood cell indices; and the presence of hyperviscosity symptoms and of bleeding diathesis, gouty arthritis, and gallstones. Assessment of hyperviscosity symptoms was based on a formal questionnaire\(^5\) that focused on the absence, presence, and degree of headaches, dizziness, blurred vision, double vision, scotoma, fatigue, myalgias, paresthesias, and depressed mentation. The New York Heart Association (NYHA) functional class was determined at the time of the most recent clinical assessment in unoperated patients and before and after any surgical intervention in patients operated on as adults. The presence of cardiac failure was based on symptoms of systemic or pulmonary venous congestion and the need for diuretics.

Twenty-five patients had transectophageal and/or transesophageal echocardiograms with color flow imaging and spectral Doppler recordings. Echocardiograms were used to semiquantify aortic regurgitation using jet width at its origin relative to the width of the left ventricular outflow tract, together with pressure half-time of the diastolic pressure difference between the aorta and the left ventricle.\(^6\,^7\)

Twenty-one patients underwent cardiac catheterization and angiography. Attention focused on the aortic-to-pulmonary collateral and biventricular flow, the native pulmoarteries, and the right and left ventricular end-diastolic pressures. The aortic-to-pulmonary collateral circulation was classified according to the recommendations of Castaneda and colleagues (Table 1).\(^8\)

Eligibility for completed hemodynamic repair was based on clinical, anatomic, and hemodynamic criteria. The anatomy and functional adequacy of native pulmonary arteries and of aortic-to-pulmonary collaterals, the pulmonary arterial pressure, biventricular function, and the degree of aortic regurgitation provided the basis for surgical selection.

In group A and B patients who had surgical interventions at age 18 years or older, data were reviewed to determine the number and nature of those interventions, the postoperative right ventricular-to-left ventricular systolic pressure ratios, and, in those who underwent completed hemodynamic repair, the operative mortality and functional class after surgery.

**Results**

Of the 785 patients referred to the UCLA Adult Congenital Heart Disease Center between 1978 and 1992, 180 were cyanotic at the time of this study. Twenty-six patients had PA-VSD, comprising 14% of the cyanotic population. Ages ranged from 18 years to 55 years with a mean ± SD of 28 ± 8.7. The age distribution is shown in Fig 1. Eighteen patients were between ages 18 and 29 years. Only two were over 40 years of age.

Fig 2 illustrates the status and outcome of all 26 cyanotic adults with PA-VSD. Sixteen survived to age 18 years without surgical intervention (group A). Ten patients were palliated before age 18 years (group B). A total of 12 patients from group A or B had surgery at age 18 years or later. Six patients died during follow-up: 5 were unoperated adult survivors who remained unoperated because of cardiac failure, and 1 was a palliated adult survivor. No patient who underwent reparative surgery beyond age 18 years has died.

**Unoperated Adult Survivors**

Table 2 summarizes the clinical data and outcome for 16 patients in group A. The mean oxygen saturation was 85% (±3.2 SD), and the mean hematocrit was 57% (±8.2 SD). Eight patients experienced mild-to-moderate symptoms of hyperviscosity. No patient was iron deficient. No patient was in NYHA class I or IV. Half the patients were in class II, and half were in class III.

Two patients endured pregnancies. At age 19, 1 patient delivered a premature but otherwise normal infant. A palliative shunt had been attempted elsewhere at age 18, but the shunt occluded within 24 hours of surgery. The patient was NYHA class II before and after pregnancy. The second patient endured three pregnancies, all ending in abortion; the first two were spontaneous, and the third was therapeutic.

Thirteen of 16 group A patients had aortic regurgitation. In 7 patients, the regurgitation was moderate or severe. The 5 group A patients who remained unoperated had cardiac failure, which was biventricular in 3 patients and right ventricular in 2. Two patients with

| Table 1. Classification of Pulmonary Blood Flow in PA-VSD |
|-----------------|---------------|--------|----------|
| Nonatretic MPA  | PDA           | RPA-LPA Confluence | APCA   |
| Type 1          | +             | +      | +        | -       |
| Type 2          | -             | +      | -        | +       |
| Type 3          | -             | -      | +        | -       |
| Type 4          | -             | -      | -        | +       |

APCA indicates aortopulmonary collateral arteries; LPA, left pulmonary artery; MPA, main pulmonary artery; PA-VSD, pulmonary atresia with ventricular septal defect; PDA, patent ductus arteriosus; and RPA, right pulmonary artery. Modified from Castaneda et al.\(^8\)
biventricular failure had moderate or severe aortic regurgitation. In the 8 patients in whom it was measured, the left ventricular end-diastolic pressure was 15 to 28 mm Hg in 4 and 10 mm Hg or less in 4. No patient with elevated left heart filling pressures had aortic regurgitation that was more than mild. Right ventricular end-diastolic pressure was 12 to 22 mm Hg in 5 and 8 mm Hg in 2 of the 7 patients in whom it was measured.

TABLE 2. Clinical Profile in Unoperated Adult Survivors (Group A)

<table>
<thead>
<tr>
<th>No. of Patients</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Female</td>
<td>16</td>
</tr>
<tr>
<td>Male</td>
<td>10</td>
</tr>
<tr>
<td>Age, y</td>
<td>32±9.5*</td>
</tr>
<tr>
<td>Oxygen saturation, %</td>
<td>85±3.2*</td>
</tr>
<tr>
<td>Hematocrit level, %</td>
<td>57±8.2*</td>
</tr>
<tr>
<td>Functional class</td>
<td></td>
</tr>
<tr>
<td>II</td>
<td>8</td>
</tr>
<tr>
<td>III</td>
<td>8</td>
</tr>
<tr>
<td>Aortic regurgitation</td>
<td></td>
</tr>
<tr>
<td>Mild</td>
<td>6</td>
</tr>
<tr>
<td>Moderate</td>
<td>5</td>
</tr>
<tr>
<td>Severe</td>
<td>2</td>
</tr>
<tr>
<td>Cardiac failure</td>
<td>5</td>
</tr>
<tr>
<td>Unoperated mortality</td>
<td>5</td>
</tr>
<tr>
<td>Considered for surgery</td>
<td>13</td>
</tr>
<tr>
<td>Pulmonary blood flow</td>
<td>10</td>
</tr>
<tr>
<td>Type 3</td>
<td>7</td>
</tr>
<tr>
<td>Type 4</td>
<td>3</td>
</tr>
<tr>
<td>Operated (after age 18 years)</td>
<td>5</td>
</tr>
</tbody>
</table>

*Values are mean±SD.

Thirteen of the 16 unoperated adult survivors were considered for surgery either by our group or by the referring physician. In 4 patients, ventricular function was deteriorating. In 1 patient, palliative surgery had been attempted before pregnancy (see above). Seven other patients experienced progressive effort dyspnea or decreasing exercise capacity. In the 10 patients who underwent cardiac catheterization, 7 had type II and 3 had type III pulmonary artery patterns (Table 1). Five patients in group A were considered eligible for surgery. Four have undergone completed hemodynamic repair with unifocalization in two or three stages. No surgical intervention was performed in patients with cardiac failure. All operated patients are alive.

Five of the 16 patients in group A have died. All of the deceased were part of the 11 patients who remained unoperated. The mean age at time of death for unoperated adult survivors was 33 years (±12.5 SD). Their cardiac status and cause of death are summarized below.

**Palliated Adult Survivors**

Table 3 summarizes the clinical findings in 10 cyanotic adults who had palliative surgical procedures before age 18 (group B). The mean systemic arterial oxygen saturation was 85% (±4.8 SD), and the mean hematocrit was 52% (±10.9 SD). Two of the 10 patients had moderate symptoms of hyperviscosity (hematocrits, 68% and 62%; iron replete).

Palliative procedures included five Blalock-Taussig shunts, four Waterston shunts, one Potts shunt, one right ventricular--to-pulmonary artery conduit without VSD closure, and four Gore-tex or venous aortic-to-pulmonary shunts. One patient had the first stage of unifocalization repair at age 17. Before age 18, 4 patients had one intervention, 3 patients had two interventions, and 3 patients had three interventions.

The majority of group B patients were in NYHA class II. Three had cardiac failure, which was left ventricular in 2 and right ventricular in 1. Of the 2 patients with left ventricular failure, 1 had severe chronic aortic regurgitation. Six other patients had mild-to-moderate aortic regurgitation. Left and right ventricular end-diastolic pressures ranged from 8 to 12 mm Hg and 7 to 14 mm Hg, respectively.

Nine of the 10 patients in group B were considered for additional surgical intervention at or beyond age 18. The indications that prompted consideration were increasing cyanosis in 2, progressive effort dyspnea in 6, and completion of unifocalization in 1. In the latter patient, hemodynamic repair had been undertaken at age 17 because of a decrease in exercise tolerance beginning at age 15. Six patients had a type III pulmonary artery pattern, 1 had type II, and 3 had type IV. Hemodynamic repairs were undertaken after age 18 in 7 patients in group B and were completed in 6, and 1 patient awaits the final stage. All operated patients beyond age 18 are alive. One patient in group B died in another hospital from acute appendicitis (see below).

**Cyanotic Adult Nonsurvivors**

Table 4 summarizes the data on 6 patients in groups A and B who survived to age 18 but died during follow-up. Five were unoperated adult survivors who remained unoperated, and 1 was a palliated adult
survivor. The mean age at death was 31 years (±12.1 SD). Four patients who died were septic. All 4 had advanced right, left, or biventricular failure. One was hospitalized elsewhere with acute appendicitis and died at age 22. He had chronic severe aortic regurgitation with a left ventricular ejection fraction of 25% at the time of his last outpatient visit. The second patient died at age 28 with acute severe aortic regurgitation caused by infective endocarditis. She sustained fatal pulmonary hemorrhage before cardiopulmonary bypass in a futile attempt at aortic valve replacement. The third patient died at age 54 with biventricular failure complicated by renal failure and Gram-negative peritonitis. She had calcific aortic stenosis and moderate aortic regurgitation. The fourth patient had severe right ventricular failure, was in NYHA class III, and died with Gram-negative sepsis associated with pulmonary and brain abscesses. The cause of death was unknown in 2 of 6 patients. Both had been assessed for surgery but were considered inoperable. One had moderate biventricular failure, moderate mitral regurgitation, and symptomatic nonsustained ventricular tachycardia. The other was 1 of the 2 pregnant patients who was last seen at the time of her therapeutic abortion, when she was clinically stable with moderate symptoms of hyperviscosity. A catheterization 1 year before her death disclosed a pulmonary artery pressure of 80/40 mm Hg, a left ventricular end-diastolic pressure of 20 mm Hg, and no detectable aortic regurgitation. In the 26 patients in groups A and B, 8 had cardiac failure, and 5 have died. Of the 3 patients with severe aortic regurgitation, 2 have died.

<table>
<thead>
<tr>
<th>TABLE 3. Clinical Profile in Palliated Adult Survivors (Group B)</th>
<th>No. of Patients</th>
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</thead>
<tbody>
<tr>
<td>Total</td>
<td>10</td>
</tr>
<tr>
<td>Female</td>
<td>7</td>
</tr>
<tr>
<td>Male</td>
<td>3</td>
</tr>
<tr>
<td>Age, y</td>
<td>21±8.3*</td>
</tr>
<tr>
<td>Oxygen saturation, %</td>
<td>85±4.8*</td>
</tr>
<tr>
<td>Hematocrit level, %</td>
<td>52±10.9*</td>
</tr>
<tr>
<td>Functional class</td>
<td></td>
</tr>
<tr>
<td>II</td>
<td>7</td>
</tr>
<tr>
<td>III</td>
<td>3</td>
</tr>
<tr>
<td>Aortic regurgitation</td>
<td></td>
</tr>
<tr>
<td>Mild</td>
<td>4</td>
</tr>
<tr>
<td>Moderate</td>
<td>2</td>
</tr>
<tr>
<td>Severe</td>
<td>1</td>
</tr>
<tr>
<td>Cardiac failure</td>
<td>3</td>
</tr>
<tr>
<td>Considered for surgery</td>
<td>9</td>
</tr>
<tr>
<td>Pulmonary artery blood flow</td>
<td></td>
</tr>
<tr>
<td>Type 2</td>
<td>1</td>
</tr>
<tr>
<td>Type 3</td>
<td>6</td>
</tr>
<tr>
<td>Type 4</td>
<td>3</td>
</tr>
<tr>
<td>Mortality</td>
<td>1</td>
</tr>
<tr>
<td>Operated (after age 18 years)</td>
<td>7</td>
</tr>
</tbody>
</table>

*Values are mean±SD.

Operated Cyanotic Adult Survivors

Table 5 summarizes the data on group A and B patients who had a surgical intervention at or beyond age 18. Mean age at intervention was 21 years (±4.1 SD) for the unoperated adult survivors and 20 years (±2.1 SD) for the palliated adult survivors. Twenty-two of the 26 patients in both groups were evaluated for palliative or reparative surgery; 12 were considered suitable candidates. The indications for intervention were increasing effort dyspnea in 10, increasing cyanosis in 2, and in anticipation of pregnancy in 1. Of 12 patients, 5 survived to age 18 without surgical intervention, and 7 had previous palliative procedures in in-

<table>
<thead>
<tr>
<th>TABLE 4. Clinical Profile in Cyanotic Adult Nonsurvivors</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total</td>
<td>6</td>
</tr>
<tr>
<td>Age at time of death, y</td>
<td>31±12.1*</td>
</tr>
<tr>
<td>Unoperated adult survivors</td>
<td>5</td>
</tr>
<tr>
<td>Palliated adult survivors</td>
<td>1</td>
</tr>
<tr>
<td>Cause of death</td>
<td></td>
</tr>
<tr>
<td>Unknown</td>
<td>2</td>
</tr>
<tr>
<td>Sepsis</td>
<td>4</td>
</tr>
<tr>
<td>Cardiac status at last assessment</td>
<td></td>
</tr>
<tr>
<td>Cardiac failure</td>
<td>5</td>
</tr>
<tr>
<td>Severe aortic regurgitation</td>
<td>2</td>
</tr>
</tbody>
</table>

*Value is mean±SD.

<table>
<thead>
<tr>
<th>TABLE 5. Cyanotic Adults Operated at ≥18 Years of Age</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total</td>
<td>12</td>
</tr>
<tr>
<td>Unoperated adult survivors</td>
<td>5</td>
</tr>
<tr>
<td>Palliated adult survivors</td>
<td>7</td>
</tr>
<tr>
<td>Age at “adult” intervention, y</td>
<td>21±3.3*</td>
</tr>
<tr>
<td>Procedures</td>
<td></td>
</tr>
<tr>
<td>Palliative</td>
<td>1</td>
</tr>
<tr>
<td>Awaiting completion of unifocalization</td>
<td>1</td>
</tr>
<tr>
<td>Completed hemodynamic repair</td>
<td>10</td>
</tr>
<tr>
<td>Mean right ventricular–to–left ventricular pressure ratio</td>
<td>0.45±0.16*</td>
</tr>
<tr>
<td>Number of interventions per patient (range)</td>
<td>1-3</td>
</tr>
<tr>
<td>Pulmonary artery blood flow</td>
<td></td>
</tr>
<tr>
<td>Type 2</td>
<td>1</td>
</tr>
<tr>
<td>Type 3</td>
<td>11</td>
</tr>
<tr>
<td>Type 4</td>
<td>1</td>
</tr>
<tr>
<td>Duration of follow-up</td>
<td>2 mo-10 y</td>
</tr>
<tr>
<td>Functional class</td>
<td></td>
</tr>
<tr>
<td>Unknown</td>
<td>2</td>
</tr>
<tr>
<td>Class I</td>
<td>8</td>
</tr>
<tr>
<td>Mortality</td>
<td>0</td>
</tr>
</tbody>
</table>

*Values are mean±SD.
fancy, childhood, or adolescence. The majority of patients who underwent surgery had a type III pulmonary artery pattern and were in NYHA class II before operation as an adult. One patient had a palliative aortic-to-pulmonary shunt in another institution at age 18, with no procedures thereafter.

A total of 10 patients have undergone completed hemodynamic repair, and 1 is awaiting completed repair. Only 1 was in cardiac failure. Four patients from group A had unifocalization in two or three stages. Seven patients in group B have had procedures, consisting of conduit insertion and VSD closure in 7, conduit replacement in 2, pulmonary artery reconstruction in 4, unifocalization in 2, and tricuspid valvuloplasty in 1. The mean±SD intraoperative right ventricular-to-left ventricular pressure ratio for the patients who underwent completed repair was 0.45±0.16 (range, 0.30 to 0.75). The duration of follow-up for the 9 patients who had completed hemodynamic repair ranged from 2 months to 10 years. Excluding the 1 patient operated on 10 years earlier, the mean duration of follow-up was 9 months (±4.6 SD). There were no reoperations after age 18 and no early or late postoperative deaths. All 10 patients are in NYHA class I. Two of 12 operated patients had moderate aortic regurgitation before hemodynamic repair. The degree of regurgitation remained stable in both patients. The one patient in whom a 10-year follow-up is available experienced progression from mild-to-moderate aortic regurgitation, a decrease in left ventricular ejection fraction to 35%, and an increase in left ventricular end-diastolic dimension to 64 mm. This patient was an unoperated adult survivor who had unifocalization in two stages at age 19.

**Discussion**

Survival of patients with PA and VSD depends on the adequacy of pulmonary blood flow derived from direct and indirect aortic-to-pulmonary collateral circulation. Natural history studies are sparse and have generally originated from large groups of patients with Fallot’s tetralogy in which age distribution was heavily weighted toward neonates, infants, and children. Between 1928 and 1967, there were six clinical or necropsy studies that comprised a total of 73 patients with PA-VSD ranging in age from 9 days to 33 years.\(^9\)-\(^\text{14}\) In three studies, the age at time of death ranged from a median of 11 months to 17 months.\(^\text{11,14,15}\) The largest single experience was a report of 782 patients with Fallot’s tetralogy, of whom 54 had PA. In this subgroup, life expectancy without surgery was 50% at age 1 year and 8% at age 10 years.\(^2\) Somerville reported on 39 patients with PA-VSD ranging in age from 2 years to 15 years, of whom only 7 were unoperated.\(^\text{16}\) Longevity beyond the third decade has been documented,\(^3\)-\(^4\) with the oldest reported survivor 54 years of age.\(^3\) None of the above studies addressed life expectancy in unoperated adult survivors. In our group of 16 patients with PA-VSD who reached age 18 without operation, 10 were in their third decade, 5 in their fourth decade, 2 survived to age 50, 1 is alive at 51, and 1 died at age 54. Almost one third of our unoperated adults and almost one half of those who remained unoperated died during follow-up at a mean age of 33 years. Unoperated PA-VSD exhibits a bimodal
mortality pattern. The majority of unoperated patients with either inadequate or excessive aortic-to-pulmonary blood flow die in infancy or childhood. A small percentage survive to adulthood because of an adequate but not excessive pulmonary circulation. However, even in these naturally selected adult survivors, longevity beyond the third decade is exceptional, and right ventricular or biventricular failure may preclude operation.

Functional capacity and symptomatic stability of unoperated adult survivors have not previously been established. Exercise testing in unoperated patients ranging in age from 4 to 27 years disclosed only 21% of predicted normal work capacity.17 None of our unoperated adult survivors were asymptomatic, and half had at least some limitation in activities of daily living. Eighty percent had been considered for surgery either by us or by a referring physician, most often by the end of the second decade of life, because of a progressive decrease in exercise capacity. This deterioration in functional capacity, which can become manifest in late childhood or adolescence, is thought to occur when pulmonary blood flow is no longer commensurate with the needs imposed by rapid growth and by the desire of older children or adolescents to increase their activity levels. Progressive stenosis of collaterals, failure of collateral growth, and an increase in pulmonary (collateral) vascular resistance18–20 are believed to account for the functional deterioration.

Cardiac failure occurred in 30% of our unoperated and palliated adult survivors. Left and right ventricular end-diastolic pressures were elevated in more than half the patients in whom it was measured. Most of these patients were clinically in heart failure, which is in agreement with observations of Sudhir et al,2 who found congestive heart failure and an elevated right or left ventricular end-diastolic pressures in half the adults with unoperated PA-VSD. Of 6 of our patients who died during follow-up at UCLA, 5 had cardiac failure at the time of their last outpatient visit. All unoperated survivors with heart failure died during follow-up. These observations underscore the need to proceed with completed repair as early as possible to decrease the incidence of ventricular dysfunction.

Survival with PA-VSD is intimately related to aortic-to-pulmonary blood flow. The oldest survivors are those with adequate but not excessive aortic-to-pulmonary collateral circulations, but the long-term price is chronic volume overload of the left ventricle. These patients have a limited cardiac reserve. Cardiac failure is a marker of poor outcome.

Aortic regurgitation of some degree occurred in 77% of our patients and was moderate or severe in 44% and 33% of unoperated and palliated adult survivors, respectively. Our oldest patient, a woman 58 years old, had aortic regurgitation and calcific aortic stenosis. In patients with Fallot's tetralogy and PA, a 14% incidence of aortic regurgitation has been reported that is three times more common than in patients with nonatretic pulmonary valves.21 Sudhir et al found that 47% of unoperated adults with Fallot's tetralogy and PA had aortic regurgitation, usually moderate. Capelli et al22 reported 18 patients with PA-VSD and aortic regurgitation, all of whom were between the ages of 12 and 42 years. The regurgitation was progressive and was believed to have been related to the mechanical effects of a dilated aortic root on the aortic ring and on malcoap-
tation of inadequately supported aortic cusps. Infec-
tive endocarditis can aggravate the regurgitation gradu-
ally or suddenly. Because the incompetent aortic valve
overrides the ventricular septum, the regurgitant flow is
biventricular. Of particular concern is regurgitation into
the right ventricle, which is already operating against
systemic resistance. The effects of closing the VSD and
thus committing the entire regurgitant volume to the
left ventricle is problematic. One patient whom we
followed for 10 years after unifocalization experienced
progression of aortic regurgitation that postoperatively
was mild with a normal left ventricular ejection fraction
and presently is moderate with dilatation of the left
ventricle and an ejection fraction of 35%. Aortic regur-
gitation is a significant hemodynamic variable in both
unoperated and palliated adult survivors with PA-VSD.
Closure of the VSD is not likely to alter the course of the
regurgitation. The abnormal aortic valve may there-
fore be an important postoperative residuum, serving as
a substrate for infective endocarditis as well as for
progressive volume overload of the left ventricle. The
aortic valve should therefore be addressed appropri-
ately at the time of the definitive surgical procedure.

The palliated adult survivor is likely to have had
inadequate pulmonary blood flow in infancy or child-
hood, thus requiring one or more shunts. The unoper-
ated adult survivor is likely to have had either adequate
or excessive pulmonary blood flow in infancy or child-
hood. These differences in anatomic and physiological
substrates as well as surgical palliation or lack thereof
contribute importantly to outcome and prognosis. Ac-
ccordingly, a valid comparison of survival patterns in
groups A and B cannot be made. However, some
conclusions can be drawn from the palliated adult
survivors as a group. The majority were in functional
class II, a third experienced cardiac failure, but mortal-
ity for the group as a whole was low. Nevertheless, 90%
of patients in this category were considered for further
intervention after age 18, usually because of progressive
cyanosis or exercise limitation. Of the patients consid-
ered for surgery, 78% were suitable candidates for
completed hemodynamic repair at a mean age of 20
years. There were no operative deaths in this group, but
the long-term postoperative fate of these patients is
unknown. In infants and children with inadequate pul-
monary blood flow, shunt palliation can prolong sur-
vival into early adulthood but does not seem to provide
an adequate long-term solution. The majority of these
patients experience a decrease in functional capacity in
late adolescence or early adulthood.

Palliative interventions may involve systemic-to-pul-
mmonary arterial shunts that can be used as the first stage

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**Fig 5.** Drawing showing definitive repair in a patient with a previous bilateral unifocal-
ization. The ventricular septal defect is
closed with a patch inserted through a right
ventriculotomy. The anastomosis between
the right unifocalization and aorta is taken
down, and the aortotomy is oversewn. The
small Gore-Tex graft that provides inflow to
the left unifocalization is ligated with surgical
cips and divided. Continuity between the
right ventriculotomy and unifocalizations is
established using a conduit composed of
Dacron proximally and a valved homograft
distally.
in unifocalization of pulmonary blood flow. Extensive, variable arborization of aortic-to-pulmonary collaterals and of the pulmonary vascular bed requires the use of synthetic conduits, pericardial tubes, or homografts to centralize pulmonary blood flow into hypoplastic branch pulmonary arteries, sometimes stimulating their growth.\textsuperscript{1,23,24} Hemodynamic repair is completed by establishing continuity between the right ventricle and the pulmonary circulation with an external valved conduit. Collateral sources of pulmonary blood flow are interrupted, and the VSD is closed (Figs 3 through 5).

Surgical management at various stages of hemodynamic repair has been reported in five series totaling 307 patients (range, 34 to 139 per study).\textsuperscript{23,25-28} Ages ranged from less than 1 year to more than 20 years with a median age of less than 10. Overall mortality rates were 13% to 45%, but the data are skewed by a preponderance of neonates, infants, and children. Reoperation rates range from 15% to 23%,\textsuperscript{23,26} Survivors of completed hemodynamic repair are described as stable, asymptomatic, or in functional class I.\textsuperscript{23,26,28} The applicability of this information to naturally selected or palliated adult survivors deserves comment. In our group of 12 repaired cyanotic adults who were either natural or palliated survivors, there were no deaths or reoperations at a mean follow-up of 9 months (±4.6 SD). The mean postoperative right ventricular-to-left ventricular pressure ratio for the 9 patients with completed hemodynamic repair was 0.45, which is appropriate for a favorable postoperative outcome. Importantly, only 1 of these 12 patients was in cardiac failure before hemodynamic repair. All repaired patients are in functional class I.

Overt postoperative electrical ventricular instability was not observed, but the duration of follow-up was relatively short. Variables that influence the risk of postoperative ventricular tachyarrhythmias include age at operation (obligatory adulthood in this study) and the function and systolic pressure in the incised right ventricle. Every attempt should be made to achieve as great a reduction in right ventricular systolic pressure as is technically feasible and to select adult patients for completed hemodynamic repair while right ventricular function is preserved.

The cyanotic, hypoxic infant or child is a significantly higher surgical risk than the relatively stable adolescent who slowly outgrows the collateral-dependent pulmonary blood flow. Cyanotic adults who experience gradual deterioration but retain satisfactory ventricular function can undergo hemodynamic repair at a low surgical risk. The high operative mortality reported when patients of all ages are collectively considered does not appear to be applicable to adults. Sufficient numbers of adult patients and sufficient durations of follow-up are not yet available to determine reoperation rates, but progressive aortic regurgitation and residual or progressive right ventricular outflow tract gradients may be important variables in postoperative adults. The use of adult-sized valved conduits can be expected to reduce the need for reoperation for conduit obstruction. The incidence of aortic regurgitation is greater in adults than in children. Accordingly, surgical intervention may require aortic valve repair or replacement.

The issue of whether aortic-to-pulmonary collateral blood flow can provide sufficient clinical stability to obviate the need for multistage unifocalization procedures has been a matter of controversy. Our data show that even in adults who survive by virtue of adequate collaterals, longevity beyond the fourth decade is exceptional, most if not all patients are symptomatic, and one third who are left unoperated or palliated develop congestive heart failure. Cyanotic survivors are usually considered for surgery in late adolescence or early adulthood because of a decrease in exercise tolerance. In our series, half were eligible for completed hemodynamic repair. Of the unoperated adult survivors who remained unoperated, almost half have died. Of those who had completed hemodynamic repair, none have died at a mean follow-up of 9 months. It appears from our observations that even apparently "stable" cyanotic adolescents are subject to deterioration by the fourth decade and that these patients represent a significantly lower reparative surgical risk than infants and children. Whether complex unifocalization techniques are destined to provide the best long-term surgical solution remains uncertain, but if left alone, these natural survivors will not do well.

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